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THE MISSING LINK FOR LUPUS ANTICOAGULANTS

For years now we have dealt with an unknown concerning the fact that the Lupus Anticoagulants are physiologically linked to thrombotic conditions and not to bleeding as could be expected from its name. The mechanism by which this autoimmune condition caused a thrombosis was always very speculative. In a recent article (1), the link has finally been described and it involves the interaction of vonWillebrand Factor (vWF) with its activation inhibitor Beta-2-Glycoprotein I.(B2GPI). B2GPI is a common circulating glycoprotein which regulates the activity of vWF to interact with platelets. Thus, if B2GPI is decreased by the presence of a Lupus Anticoagulant antibody, vWF will bind to platelets and cause clumping. The article describes how vWF is produced in high molecular weight forms by the endothelium and how it's broken down by ADAMTS13 and circulates as a globular complex. This globular complex does not interact with platelets unless it is exposed to high shear in the circulation which opens the complex and allows the vWF to interact with the platelet glycoprotein (Gp)-Ib/IX/V and form platelet clumps. Therefore, our knowledge of deficiencies of ADAMTS13 which leads to Thrombotic Thrombocytopenia(TTP) due to high molecular weight forms of vWF binding to platelets, is now enhanced by how normal molecular weight forms can be activated or induced to bind to platelets and cause thrombosis. This also fits with the finding that certain forms of Lupus Anticoagulants that are associated with a high titer anti-B2-glycoprotein I antibody, are 20 fold more likely to cause thrombosis or be associated with spontaneous abortions than Lupus Anticoagulants which are linked to Prothrombin antibodies. This article provides exciting reading combing the knowledge of Lupus Anticoagulants, vWF disease (specifically Type IIB) and platelet interactions.

- (1) Regulation of von Willebrand factor-platelet interactions. Lenting, P.J., Pegon, J.N., Groot, E., de Groot, P.G.. Thrombosis and Haemostasis 104. 3, 2010 p449-454. Site reference:
<http://www.schattauer.de/en/magazine/subject-areas/journals-a-z/thrombosis-and-haemostasis/issue/special/manuscript/13198/show.html>

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